

Silver Linings: The Experience of Hope in Dyads with Motor Neuron Disease. An Explorative Study

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Abstract

Objective: Research on the role of hope in coping with fatal Motor Neuron Disease including Amyotrophic Lateral Sclerosis (ALS) and Progressive Muscular Atrophy (PMA) is scarce. In this study we aimed to enhance our understanding of experienced hope in dyads diagnosed with Motor Neuron Disease (MND).

Methods: Eligible purposively sampled MND patients attending four large specialized MND care teams in The Netherlands were approached to participate in this study. Consenting patients invited an informal caregiver of their choice to participate in the interviews. The appointed caregivers all appeared to be Family Caregivers (FCs). Qualitative semi-structured interviews were conducted in dyads of 22 MND patients and 22 Family Caregivers (FC) and thematically analyzed.

Results: Most patients and their family caregiver (dyads) experienced hope and valued it as essential for continuing living with MND after the diagnosis. Three overarching themes emerged from the interviews: hope for more life years, living meaningful and a dignified death. Dyads reported a quick shift after diagnosis from hope for a cure to hope for a meaningful remaining time. Retaining hope during disease progression meant that patients actively re-appraised and revalued their life as a buffer against distress and despair. Hopefulness of the patient with MND was an important source of hope for all family caregivers.

Conclusion: Hope is a salient and dynamic concept that plays a central role in the process of coping with MND and its devastating consequences. In the process of finding and retaining hope, patients and FCs are challenged to actively search for their inner strength, evaluate and re-evaluate what is important to them, while facing severe physical deterioration and imminent death. Hope appears an essential emotion-regulating coping-mechanism and denotes a process that by redefining and pursuing attainable goals, positive emotions are generated.

Professionals need to be aware of the fact that both patients and FCs experience and value hope in the process of accepting and living with the consequences of MND. Hope does not imply denial of the medical reality where there is no cure, but rather signify a need for and motivation to make meaning of remaining time living with MND. Hope serves as an inner force and transcends through the course of the disease and its profound psychosocial challenges. It is imperative that health professionals understand and appreciate hope from the patients' and caregivers' perspective and offer assistance where possible in achieving what is meaningful. Discussing hope and offering guidance constitutes an essential part of multidisciplinary care for persons with MND and their caregivers throughout the illness process.

Keywords Hope; Amyotrophic lateral sclerosis; Progressive muscular sclerosis; Motor neuron disease; Dyads; Caregivers; Qualitative research

Introduction

Amyotrophic Lateral Sclerosis (ALS) and Progressive Muscular Atrophy (PMA), collectively referred to as Motor Neuron Disease

(MND), are fatal neurodegenerative diseases with a rapid course. There is no proven curative treatment available. MND leads to progressive muscle weakness and stiffness, limitations in mobility, impaired speech, difficulty swallowing and eventually respiratory problems. Most patients with MND die from respiratory failure. Median survival time from onset of symptoms is three years [1]. Receiving the diagnosis of MND understandably leads to an existential shock in patients and their caregivers, forcing them to re-appraise their lives [2].

Psychological reactions to the diagnosis can range from grief and anger to a desire for hastened death [3,4]. To date, findings on the prevalence of psychological problems are inconsistent but it would appear safe to conclude that only a minority of patients suffer from clinical depression and anxiety, and that most persons with MND while having end of life concerns find a way to cope with the diagnosis and show psychological resilience [5,6]. In this context feelings of hope may play a role as demonstrated in patients with other terminal illnesses [7,8]. Herth defined hope as “*an inner power that facilitates the transcendence of the present situation and movement toward new awareness and enrichment of being*”. Indeed, hopefulness has been identified as a central need of terminally ill patients and their caregivers [9-12]. To date, research in MND on the topic of hope is for the majority limited to the perspective of patients. A few studies on hope in MND have been conducted [13-16], confirming the relevance of the concept and the need for care professionals to take time to explore hope fostering approaches in their patients. Furthering our understanding of hope in dyads with MND could help enrich the communication between patients, caregivers and professionals, and contribute to further improvement of supportive care as found in other terminal illnesses [17,18].

The aim of this qualitative study is to explore if and how hope is experienced in dyads of MND patients and their caregivers at different stages of the disease.

Methods

Participants

MND patients attending four large specialized MND multidisciplinary care teams in the Netherlands were approached to

participate in this study. Patients inclusion criteria were:- 1) Having a diagnosis of ALS or PMA according to El Escorial criteria [19] and 2) being able to communicate in Dutch. An associated diagnosis of fronto-temporal dementia (FTD) was an exclusion criterion. Patients were sampled purposively to capture diversity in: gender, age, diagnosis, first symptoms, marital situation, religion and cultural background. Additionally, patients in different illness phases were selected; 1) ‘early MND’, from four weeks after diagnosis up till one year in MND care; 2) ‘later MND’, defined as more than one year after onset of MND care; 3) ‘end of life’, as determined by the physician of the MND care team and generally considered to be the last months of life. Patients invited an informal caregiver of their choice to participate in the interviews. Inclusion criteria for the caregivers were 1) 18 years or older; 2) Able to communicate in Dutch, and 3) Invited by the recruited patient. The appointed caregivers all appeared to be Family Caregivers (FCs). Recruitment of patients continued until data saturation was reached i.e. no new themes emerged from the interviews.

Twenty-eight eligible patients were invited to participate. Of these, 2 considered themselves too emotionally unstable to participate, 2 experienced severe physical deterioration, and 2 FCs were not willing to participate, leaving 22 participating dyads in the study. The Medical Ethical Review Committees of the 4 participating centers approved the study. Written informed consent was obtained from all participants.

Interviews

A semi-structured interview was developed by our team, using open-ended questions aimed to explore patients’ and FCs views on and experiences with hope. The interviews for patients and caregivers were guided by a topic list informed by clinical experience, literature and was pilot tested Table 1.

Topic list for the patient	Topic list for the caregiver
Introduction of the interviewer and general information about the interview	Introduction of the interviewer and general information
Life with ALS or PMA	Life with ALS or PMA
- history of ALS/PMA	- ALS/ PMA impact on life
Hope and its value	Hope and its value
- meaning of the word hope	- meaning of the word hope
- hope before the diagnosis	- hope before the diagnosis
- hope in this situation	- hope in this situation
- value of hope	- value of hope
Content of hope	Content of hope
- what to hope for	- what to hope for
Retaining hope	Retaining hope
- strengthening hope	- strengthening hope
- threat to hope	- threat to hope
Selection of the significant other	Caregiver’ view on hope of the patient
Patients view on hope of significant other	Conclusion and follow up procedure including member check

Conclusion and follow up procedure including member check	
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Table 1: Topic list for patient and caregiver

All interviews were conducted by a psychotherapist involved in care for MND patients (SdM), and a research psychologist (LNCV). Interviews with patients and their FCs were conducted separately if possible. All interviews were audio- taped and transcribed verbatim.

Data analysis

Analyses were performed in parallel with the interviewing, in a multiphase process, following guidelines for qualitative research and thematic analysis [20-22] and using MAXqda2 software [23]. Basic coding schemes, based upon the two pilot interviews, were developed and in agreement defined for the interviews with patients and FCs, respectively, by the interviewers (SdM, LNCV) to optimize content validity. The same interviewers coded the interviews independently. After each interview any discrepancies in thematic coding were thoroughly discussed, until consensus was reached. The coding schemes were revised based on the results of the independent interim analyses by the interviewers. All interviews were coded according to

the final coding scheme. The first 22 interviews (14 patients, 9 FCs) were independently coded, showing almost full consensus. The remaining 19 interviews were coded by the first author only. The accuracy of the content of a sample of 14 interview summaries was ('member') checked by patients and their FCs. Codes were grouped together into categories and core themes were derived. Multiple senior researchers (AB, EMS, HH) critically reviewed primary documents, coding schemes, interpretations and definition of themes as a quality check. Quotes illustrating core components of the emerging themes were carefully selected as shown appendix 1.

Results

Participant characteristics

Participant characteristics are summarized in Table 2.

Illness phase (a)	Participant	Sex (b)	Age	Cultural back ground	Marital situation (c)	Diagnosis	First symptom	Speech ability	Daily use wheelchair	NIV (e)	Presence during interview (f)	Relation FC	Sex FC	Age FC
1	1	m	62	Aruba	single	ALS	spin/bul	normal	no	no	separate	sister	f	68
	4	m	69	Suriname	couple a.ch	ALS	bulbar	limited	yes	yes	separate	spouse	f	55
	5	m	48	Netherlands	family	PMA	spinal	normal	no	no	separate	spouse	f	44
	6	m	54	Netherlands	family	ALS	bulbar	normal	no	no	separate	spouse	f	49
	8	f	78	Israel	single	PMA	spinal	normal	no	no	present	daughter	f	43
2	3	m	73	Netherlands	couple a.ch	ALS	spinal	normal		no	Present	spouse	f	71
	9	m	47	Netherlands	couple n.ch	PMA	spinal	no	yes	no	mixed	spouse	f	53
	10	f	54	Yugoslavia	couple n.ch	ALS	spinal	limited	yes	no	separate	spouse	m	53
	11	m	51	Netherlands	family	ALS	spinal	normal	no	no	mixed	spouse	f	48
	14	m	62	Netherlands	couple a.ch	ALS	bulbar	normal	yes	yes	present	spouse	f	61
	15	m	74	Netherlands	couple a.ch	PMA	spinal	normal	yes	no	separate	spouse	f	80
	16	m	57	Netherlands	couple a.ch	ALS	spinal	limited	yes	yes	present	spouse	f	54
	17	m	76	Netherlands	widower	PMA	spinal	normal	no	no	present	daughter	f	45
	19	f	72	Indonesia	widow	ALS	spinal	normal	yes	no	present	sister	f	73
	20	m	54	Netherlands	family	ALS	spinal	normal	no	no	separate	spouse	f	51
	21	f	56	Netherlands	couple a.ch	ALS	spinal	normal	yes	no	present	spouse	m	55
	22	m	37	Netherlands	family	ALS	spinal	normal	yes	no	separate	spouse	f	38
	23	f	26	Netherlands	couple n.ch	ALS	spinal	normal	no	no	present	spouse	m	31

	24	f	52	Netherlands	family	ALS	bulbar	no	yes	no	present	spouse	m	52
3	12	m	69	Netherlands	couple a.ch	ALS	spin/bul	no	yes	yes	mixed	spouse	f	47
	13	m	78	Netherlands	couple a.ch	PMA	spinal	limited	yes	yes	separate	spouse	f	67
	18	f	46	Netherlands	couple n.ch	ALS	spinal	limited	yes	yes	separate	spouse	m	48
<p>Note: (a) 1=early MND care: between 4 weeks and 1 year after diagnosis MND/2=later MND care: ≥ one year after diagnosis MND/3=end of life (b) f=female/m=male (c) family=children living with their parent/Couple n.ch= couple with no children/couple a.ch= couple with adult children not living with their parents (d) spin/bul=first symptoms were a combination of spinal and bulbar symptoms/bulbar=first symptoms were exclusively bulbar/spinal=first symptoms were exclusively spinal. (e) NIV=Non-invasive Ventilation (f) present=the interview with the patient was conducted in the presence of the significant other (and vice versa)/separate=the interview with the patient was conducted in the absence of the significant other i.e. not in the same room (and vice versa)/mixed=the interview with the patient and significant other was recorded as one interview.</p>														

Table 2: Characteristics of participants.

In total 22 patients (15 male, 7 female) and 22 FCs (18 spouses, 2 daughters and 2 sisters) were interviewed. Mean age of the patients and FCs was 59 (range 26-78) and 54 (range 31-80) respectively. Sixteen patients were diagnosed with ALS and 6 with PMA, 2 with both spinal and bulbar onset, 4 with bulbar onset and 16 with spinal as first symptoms. We included 5 patients in early MND phase, 14 patients in later MND phase and 3 patients in the end of life phase. Four patients had a non-European cultural background.

Eight patients and their FCs were interviewed separately. The other 14 patients were interviewed in the presence of their FC, either because they needed physical assistance, or on their request as they were interested in hearing each other's story. Of the 22 FCs, 19 were interviewed individually, 3 FCs served as the spokesperson, due to severe speech problems of the patients. Their stories were recorded in one interview ("mixed" in Table 2).

Participants were asked to describe whether they experienced hope, and if so, to elaborate on the content, value and meaning of hope and changes in therein during the course of MND. Participants not reporting hope were invited to elucidate their experience.

To hope or not

Patients as well as FCs confirmed to have retained feelings of hope after the first shock of receiving the diagnosis of ALS or PMA. Hope was valued as important and in fact a necessity to help them continue their lives. It provided them with something 'to hold on to', inner strength and a perspective during the illness process. Hope was valued as important in the process of regulating negative emotions.

'Yes, the courage to live comes from the things I hope for' #4P

'Hope is the last thing you've got,Yes, if you have no hope left, you might as well stop living' #9FC

Few patients and some FCs denied experiencing hope. Those who did explained that to them the term 'hope' was associated with a cure and therefore considered not realistic. However, to them not having hope was not to be mistaken for hopelessness or despair.

'There is no hope....No cure and therefore you should not focus on hope. I don't feel the need for hope, I want to face the facts' #11P

In some dyads, patient and FCs differed in their perspective on hope, which was not necessarily problematic. Interestingly, if the patient and the FC held different views concerning hope, it was always the FC who reported to have less hope than the patient. On the other hand, FCs considered the patients' hope critical for the patients' ability to carry on and maintain emotional balance. All FCs brought forward that seeing the patient effectively cope with the illness was the main source of retaining hope for themselves.

'He gives me hope. He is ill but when he is feeling optimistic and coping well, that helps me to cope, that gives me strength, because you are in this together. You need each other to be hopeful' #1FC

'Most of my hope I get from his positivity' #22FC

The content of hope

Patients and FCs emphasized that they were fully aware of the devastating medical reality and that the focus soon had shifted from hope for a cure to other, attainable things to hope for.

Three main, partly overlapping themes concerning the content of patients' and FCs' hope emerged from the interviews: a) hope for a longer life, b) hope to continue living meaningfully (pertaining to physical, psychological, behavioral and social themes), and c) hope for a dignified death and a good life for those left behind as shown in Table 3 for an overview of the different themes.

1. Hope concerning a longer life span	- for a misdiagnosis	- for a misdiagnosis
	- to living longer than prognosticated	- to living longer than prognosticated
	- for the discovery of a medicine	- for the discovery of a treatment
	- that behavior and spirit will slow down the illness process	- that behavior and spirit will slow down the illness process

	- to be with loved ones as long as possible*	- to keep the beloved patients long as possible in their midst*
2. Hope concerning living meaningful	Physical functioning themes	Physical functioning themes
	- that trial-participation will have a positive impact on the illness process	-for delay in functional loss
	- for delay in functional loss	-for stabilization of the current status of physical functioning*
	- for stabilization of the current status of physical functioning*	-for outliving the patient
	Psychological themes	Psychological themes
	-to keep autonomy	-to be able to give her/him(i.e., patient) an enjoyable life as much as possible and to live positively
	-to keep up the strength to cope well	-that the patient will have the strength to cope
	-to be mentally well prepared for the next phase or deterioration	
	- to be able to enjoy life as much as possible and to live positively	
	Behavioral themes	Behavioral themes
	-to be able to perform specific enjoyable things	-to be able to perform specific enjoyable things together*
	-for the possibility to keep experiencing interesting things	-for the possibility to keep experiencing interesting things
	-for the ordinary daily things (nice weather, to be able to go outside, a cosy home, a beautiful garden, more nice days to come)*	
	Altruistic /Social themes	Social themes
	-that trial-participation will help future patients	-to be able to continue to work
-that loved ones will get time to get used to the situation and adjust slowly, so everyone will get through the whole illness process together in a good way	-for leniency from employer	
	-for support from professionals	
3. Hope concerning a dignified death and thereafter	-on an afterlife with Christ, to find shelter in Christ's love	-to be able to cope after the patient's death
	-to leave a legacy	
	-that loved ones will be able to cope after death	Altruistic themes
	-to find the courage to speak up when the suffering is too much to bear	-that the patient will die in peace without much suffering (i.e., dying with dignity)
	-to be able to die in peace without suffering	-that the patient will pass away in a specific way

Table 3: Content of hope of patients; Content of hope of care givers

Patients hoped for slow progression of the disease, allowing them to live as long as possible with their loved ones, and being able to continue 'normal' daily activities as long as possible.

'For me, hope is that I will be able to carry on this year in a decent way, so I can live the last part of my life with dignity, With the focus on the usual things in daily life, that you normally don't think about, but now finding these actually quite important'. #5P

Likewise, FCs most frequently mentioned the hope for a slowly progressing form of the illness so they could spend more time together so to be able to experience special moments with ones partner.

'After a while, it turned out to be the slow disease type. And at that point I started hoping again and it remained. That's the only hope you

have. And for the rest? You hope you can make something of the time you have together'.#21FC

FCs felt a responsibility to help their partner achieve an enjoyable life as much as possible and hoped they would be able to fulfill that supportive role. Also, FCs who had a job hoped to be able to continue to go to work as a distraction from all the concerns and caring; they hoped for understanding and leniency from their employer.

'And luckily I find distraction in my work and nice colleagues. They all know about ALS.....I'll be honest, I'm glad I still have my work to go to and I hope I can keep doing my work even though it's just a few hours a week. Just so I can go out. I hope my boss will keep saying: well J., you can come to work any time you like, even for just a couple of hours'. FC#20

Retaining hope

Patients and FCs reported that personal and meaningful issues that were hoped for are re-appraised in the course of the disease process. In the early MND phase, shortly after diagnosis, it was found hard to experience hope. Some had difficulty grasping the truth and initially hoped for a diagnosis of another, curable or at least treatable condition.

'Earlier I had received the diagnosis cancer which later appeared to be a mistake. Then they told me I had ALS, so I still had a little hope it could be something else.' #4P

Retaining hope is not an easy or self-evident process, exemplified by three patients who reported acute depression, anxiety and even a psychosis after receiving the diagnosis. It took these patients several months to adjust. Two of them received professional psychological help. All three had managed eventually to restore their emotional equilibrium and experience feelings of hope.

In the early MND phase patients and FCs are challenged to face the 'medical truth' of having a progressive, fatal disease with a short life expectancy. In time hope transcends from hope for a cure to hope for a longer life. For some, participating in medical trials was reported as a way of actively contributing to retaining hope for a longer life—next to altruistic reasons, to help a next generation of MND patients.

Patients and FCs explained that to retain hope and to avoid becoming overwhelmed by negative emotions, continued efforts are needed throughout the disease process. A frequently mentioned way of retaining hope by patients was trying to 'live in the moment' and avoid thinking about the inevitable future.

'I'll take it as it comes, I don't think about tomorrow too much, I will take it day by day' #5P

FCs expressed concerns about the future and found it difficult to adopt a day-by-day mindset and retain hope.

'We know it will be worse and more intense in the future. But that is not for now. And that is the hope we've got: That the bad period will be delayed for a long time.' #20FC

Some patients tried to protect themselves from being overwhelmed by avoiding medical information around MND in order to preserve hope. Other protective strategies reported by patients were avoiding funerals and thinking of people who are worse off ('downward comparison').

One of the examples comes from an ALS patient whose mother had died from the same disease:

"In a way we are lucky because my mother had ALS too, so we know what to expect. It must be horrible not knowing what to expect." #20P

In the later MND phase patients' and FCs' hope mainly concerns quality of life i.e. living in a meaningful way. This also reflects hope for sufficient inner strength to cope with the relentlessly progressing physical deterioration and dependency on others and assistive devices.

Exerting emotional and cognitive control as a way to preserve hope was reflected in the report of one patient who was dependent on medical aids. She and her husband stored everything in closets and drawers, to be taken out only when needed.

Hope was experienced as fragile. This became particularly manifest when confronted with physical deterioration, especially when problems occurred with the provision of assistive devices and home modifications.

'Securing the supply of medical aids takes way too much time, often the delivery is too late, as the illness progresses fast. It is terrible to realize that if it had come in time I would have been able to use my computer longer. It is hard for me not to be able to do it myself anymore, that diminishes my hope' #10P

Patients and FCs in the end of life phase (three dyads) particularly focused their hope on a dignified death and a good life thereafter for those left behind. Some patients, also earlier in the disease process, expressed the hope that they would be strong enough to 'say stop' and discuss assisted life ending if not able to cope any longer. Of the 22 patients interviewed, 12 spontaneously mentioned to have discussed the opportunity of euthanasia with their physician already early in the disease process and had reached agreement on the procedure if the situation would arise.

FCs shared the hope for a death without suffering, as expressed by one partner who was interviewed separately from the patient: *"This morning I woke up, I really thought that he had passed away in his sleep. That is what I hope for his sake and maybe also for mine"* #15FC

Discussion

This explorative study confirms that patients diagnosed with MND and their FCs experience and value hope, while being fully aware of having a progressive, incurable disease. Hope is valued as essential to continue living with the devastating consequences of MND. In line with Soundy and Condon [15], we found MND patients and their FCs to actively use hope enabling strategies to manage their situation on a daily basis and preserve emotional wellbeing.

With regard to the content of hope, three key themes emerged from the interviews, partly in parallel with the disease trajectory: After having lost hope for a cure or treatment to halt the disease progression, hope shifts to hope for more life years (slow disease progression), living a meaningful life, and eventually hope for a dignified death.

Retaining hope does not come easy. In the process of finding and retaining hope, patients and FCs are challenged to actively search for their inner strength, evaluate and re-evaluate what is important to them, while facing severe physical deterioration and imminent death. Hope appears an essential emotion-regulating coping-mechanism and denotes a process that by redefining and pursuing attainable goals, positive emotions are generated [7,9,15,24]. As pointed out by Fanos et al. [16], it is through hope and self-transcendence that ALS patients are able to bear the seemingly unbearable. Importantly, the few patients in our study who said not to experience hope were not in despair. These patients too were actively working towards what seems central in the palliative phase i.e. maintaining emotional stability albeit by employing a different ('rational') coping strategy.

By interviewing dyads we were able to gain insight into the dynamics of hope within the relationship of patients and their caregivers. The patient's hopefulness was an important source of hope for the FCs and highly valued to help maintain a positive outlook. FCs generally expressed less hope than patients and face an ongoing challenge to live 'in the moment' while their situation as career becomes increasingly demanding and uncertain with regard to their own future [9].

There are some limitations in our study. Firstly, patients were recruited by their MND care physician, which may have resulted in a selective sample, despite the specific profiles we asked for. We did include a heterogeneous sample, although we had only three patients

in the end-of life phase. However, it is difficult to adequately predict life expectancy in MND as illustrated by the fact that some patients in the late MND phase died shortly after the interview.

Secondly, the fact that caregiving family members were invited to participate via the patients may have influenced their responses. However, we have no indication that they felt restricted. Indeed, some patients and their FCs had clearly expressed conflicting views on hope, in each other's presence.

Thirdly, we used a cross-sectional design, because it is very difficult to carry out a prospective study in this kind of pathology and therefore we were not able to investigate changes in hope prospectively. However, by purposive sampling we did gain insight into role of hope during three phases of MND care that we distinguished a priori. Evidence based reliable prediction models for illness progress in MND may become available in the near future [25].

Fourthly, we should take note of the fact that this study was performed in the Netherlands may limit external validity, i.e. generalizability. In the Netherlands, regional multidisciplinary MND care teams are in place and care is fully reimbursed, which is not the case in all countries. Moreover, it is policy for Dutch MND care physicians to openly discuss end of life issues and the wish for hastened death, in accordance with the European consensus [26]. The option of euthanasia may impact hope for a dignified death. Further cross-national research can help to establish the role of discussing hastened death in the experience of hope. Also, religion was rarely mentioned in our study as a factor fostering hope, in contrast to previous studies performed in USA, Finland, Canada and Singapore [9,27-29] and warrants further research.

Conclusion

Hope clearly is a salient concept in the context of MND care. While this study did not explicitly focus on the role of health care professionals, there are some insights that should prove helpful in supporting MND patients and their significant others. Professionals need to be aware of the fact that both patients and FCs experience and value hope in the process of accepting and living with the consequences of MND. Hope does not imply denial of the medical reality where there is no cure. Rather, hope serves as an inner force and transcends through the course of the disease and its profound psychosocial challenges. It is imperative that health professionals understand and appreciate hope from the patients' and caregivers' perspective and offer assistance where possible in achieving what is meaningful. Discussing hope and offering guidance constitutes an essential part of multidisciplinary care for persons with MND and their caregivers throughout the illness process.

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All authors have approved the final article.

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